

CASE 9

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This 6-month-old boy was the product of an uneventful pregnancy and full term, low forceps delivery. Birth weight was 5 lb. 4 oz. Jaundice was noted immediately after birth and persisted for 2 months. At 4 months his appearance was that of a day old infant. He did not respond to sound or bright objects but did respond to touch. He did not turn over. At the age of 5 months his motor development was at the 12 week level. In all other areas he functioned between the 4 and 8 week level. He died of bronchopneumonia with a temperature rising to 105° F.

At autopsy the body was 22 inches in length and weighed 7-1/4 lbs. The head circumference was about 12 inches. Gross abnormalities included bilateral bronchopneumonia and scattered foci of fatty metamorphosis of the liver. The fontanelles were small. The dura was firmly adherent to the inner table of the skull. The brain weighed 200 gm. All its components were reduced in size, but particularly the cerebral and cerebellar central white matter, striated portion of the pons and the pyramids. The ventricular system showed moderate diffuse dilatation. The lining of the lateral ventricles was diffusely thickened and mottled with tiny grayish white elevations. The central white matter in the frontal lobes was soft and grayish and contained a few tiny chalk white foci. A few similar foci were found in the subependymal region of the lateral ventricles. Large cerebral gyri were incompletely subdivided into tiny complex convolutions. In the cerebellar folia, the multiple sulci effected more complete subdivisions.